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АКТУАЛЬНІ ПИТАННЯ ТЕРЕТИЧНОЇ ТА ПРАКТИЧНОЇ МЕДИЦИНИ

Topical Issues of Clinical and Theoretical Medicine

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VERY RARE CASE OF PERSISTENT NEUTROPENIA

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In the Haematology Department of Sumy Regional Child Clinic Hospital, there was a child with the syndrome of persistent neutropenia.

This is a 13 month old female who is referred for cold symptoms, fever (T 38.5o C) and a persistently low absolute neutrophil count (ANC = WBC x (segs% + bands%)) of 40 (WBC 4.0, 1% bands, 0% segs). Her ESR is 12. She was hospitalized three weeks ago for a pseudomonas external otitis media and neutropenia that was treated with two weeks of intravenous antibiotics. Three weeks ago, her ANC was 342, with a low of 54, and at discharge her ANC was 896 (one week ago). She had been doing well since discharge until her current illness.

Of note, a CBC at birth and at 10 months of age demonstrated normal absolute neutrophil counts. There is no history of increased bacterial or fungal infections. There is no family history of recurrent bacterial infection, neutropenia, immunodeficiency disease, autoimmune disease, or malignancy. There is no history of infant deaths in the family. There has been no history of recent medication use.

Exam: VS are normal. Height and weight are at the 75th percentile for age. Physical examination had no unusual findings. No oral thrush, lymphadenopathy, hepatosplenomegaly, or skin lesions are noted.

She is hospitalized for IV antibiotics (ceftazidime). Blood cultures return negative and she is discharged after 3 days. During this hospital stay she does well. Antineutrophil antibody testing is sent off to a specialized reference laboratory (Kyiv) and it returns positive. A bone marrow examination is done (mostly because of parental concern) which shows a normal cellular marrow. During subsequent febrile illnesses, she does well clinically. Three months later, after she initially presented with neutropenia, her ANC improves to 2400. Two months later, it remains over 2000.

Diagnosis: Chronic benign neutropenia of infancy and childhood.

Neutropenia is defined by an absolute neutrophil count (ANC) <1,500/cubic mm. ANC = (%bands + %mature neutrophils) X total WBC count. Decreased neutrophil production, storage, or release; redistribution from circulating to marginated pools; or increased destruction explains most cases of neutropenia. The key determinants of infection risk are the adequacy of the bone marrow storage or reserve pool and the general robustness of the immune response. These determinants affect the ability to deliver neutrophils to infected sites and the ability of the immune system to compensate for quantitative deficiencies in neutrophils.

Neutropenia discovered during the evaluation of infection is generally a secondary finding and characterizes the general low risk of infection associated with a normal marrow reserve and immune system.

Some of the causes of neutropenia are summarized below: Kostman syndrome: primary decrease in bone marrow reserve,AR (autosomal recessive), AD (autosomal dominant), S (sex linkedrecessive), extremely rare,severe neutropenia in the newborn; cyclic neutropenia: primary cyclic (every 21 days) variations in bone marrow reserve, AD, S, 1-2 per million, regularly recurring fever every 21 days with oropharyngeal and skin infections, diagnose with CBCs 2-3 times per week for 8 weeks; nutritional: protein-calorie malnutrition, B12 deficiency, copper deficiency can result in ineffective myelopoiesis, treat by correcting deficiency; viral infection: bone marrow suppression from a direct effect of infecting virus or through an immune mechanism; chronic benign neutropenia of infancy and childhood: normal bone marrow reserve, generally thought to be mediated through an anti-neutrophil antibody, common, median age of detection 8 months, 90% detected before 14 months, most resolve spontaneously within months of diagnosis, no significant propensity to infection, treatment is supportive; autoimmune neutropenia: generally with normal bone marrow reserve but may be associated with a late maturational arrest, antibody mediated destruction of neutrophils, may have an associated primary autoimmune disorder; alloimmune neutropenia: normal bone marrow reserve, secondary to maternal anti-neutrophil antibody that has crossed the placenta,

resolves by 3-4 months of age, generally supportive care; drug-induced neutropenia: normal bone marrow reserve, but has been associated with a late maturational arrest, antibody or complement mediated neutrophil destruction, treatment consists of stopping unnecessary medications; infection related neutropenia: normal bone marrow reserve, virus induced anti-neutrophil antibody; parvovirus B19 and HIV can be screened, no treatment generally necessary; hypersplenism: normal bone marrow reserve, sequestration/possible destruction of neutrophils in the spleen, associated with malaria, TB, neoplasm, collagen-vascular diseases, hemolytic anemia, spherocytes and tailed RBC on blood smear, treat underlying disorder; pseudo-neutropenia (severe infection): normal bone marrow reserve; generally associated with increases in marginated and tissue pools, mild and spontaneously resolves; Shwachman-Diamond Syndrome: primary decrease in bone marrow reserve, AR, S, extremely rare, steatorrhea from exocrine pancreatic deficiency, metaphyseal dysplasia, 50% survival; 1/3 progress to myelodysplastic syndrome or acute myeloid leukemia, normal sweat chloride; chemotherapy: direct toxicity to neutrophil precursors results in a severe reduction in bone marrow reserve (severity dependent on the intensity of chemotherapy agents used), generally a high risk of infection with poor marrow reserve and generalized suppression of the immune system.

CONTENT OF COBALT IN BIOLOGICAL FLUIDSOFFULL-TERM NEWBORNS AS THE PREDICTOR OF PERINATAL HYPOXIC DAMAGE OF CNS

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Influence of Co content on the system mother-placenta-fetus-newborn children born with hypoxia. Determination of microelement (ME) performed in serum and erythrocytes 30 pregnant women and their infants who have suffered asphyxia at birth. Comparison group consisted of 30 healthy women and their 30 healthy full-term infants. The content of trace elements in biomaterials newborns and their mothers was determined by atomic absorption spectrophotometry mass spectrophotometer C-115M1, manufacturing NPO "Selmi" (Ukraine).

In the placenta of women who gave birth to children with hypoxia observed deficit Co, which creates conditions for faster penetration him to the fetus, but at the same time accumulation feature is suppressed. Serum and red blood cells of pregnant women who gave birth to children with CNS observed a significant lack of cobalt. We also investigated the features of ME content in serum and erythrocytes and especially their renal excretion in term neonates who underwent perinatal hypoxic of the CNS. These children in neonatal there a significant imbalance of serum and erythrocyte content Co. One of the factors of this imbalance ME if perinatal hypoxic of the CNS in term infants is inhibition of excretion of ME. It is proved that an imbalance of serum and erythrocyte Co content in the body of the fetus and newborn, resulting in reduction of Co pregnant women and dysfunction placental hypoxia. Predictors properties of Co detected in serum ≥3,01 mmol/l in erythrocytes ≥0,61 mg / mg ash and urine ≤0,40 mmol / l, respectively. Prognostic significance (index of informing and prognostic factor) was high.

THE VIOLATIONS OF NEURO-MENTAL DEVELOPMENT OF CHILDREN WHO HAVE SUFFERED FROM PERINATAL HYPOXIC DAMAGE OF CENTRAL NERVOUS SYSTEM

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The problem of children adaptation who suffered from perinatal hypoxic damage (PHD) of the central nervous system (CNS) is very important. The brain damage on the early stages of ontogeny violates deterministic evolutionary scenario of pre-, intra-and postnatal adaptation, slows ripening of parameters of functional CNS, which increase the likelihood of developing of secondary cerebral defects. The effects of PHD of CNS reflect not only the severity of injuries, but the effectivenes of